SANTA CRUZ BIOTECHNOLOGY, INC.

# HCCS (D-12): sc-168060

## BACKGROUND

HCCS (holocytochrome c-type synthase), also known as CCHL (cytochrome c-type heme lyase), is a 268 amino acid mitochondrial inner membrane protein that belongs to the cytochrome c-type heme lyase family. Containing two HRM (heme regulatory motif) repeats, HCCS participates in the covalent linkage of a heme group to an apoprotein of cytochrome c. The gene encoding HCCS maps to the human X chromosome. Defects to this gene cause microphthalmia syndromic type 7 (MCOPS7), also known as MIDAS syndrome or microphthalmia with linear skin defects (MLS). MCOPS7 is an X-linked male-lethal disorder that results in eye deformation, unilateral or bilateral microphthalmia, linear skin defects in affected females, and in utero lethality for males.

## REFERENCES


1. Schaefer, L., et al. 1996. Cloning and characterization of a putative human holocytochrome c-type synthetase gene (HCCS) isolated from the critical region for microphthalmia with linear skin defects (MLS). Genomics 34: 166-172.
2. Van den Veyver, I.B., et al. 1998. Genomic structure of a human holocytochrome c-type synthetase gene in Xp22.3 and mutation analysis in patients with Rett syndrome. Am. J. Med. Genet. 78: 179-181.
3. Prakash, S.K., et al. 2002. Loss of holocytochrome c-type synthetase causes the male lethality of X-linked dominant microphthalmia with linear skin defects (MLS) syndrome. Hum. Mol. Genet. 11: 3237-3248.
4. Wimplinger, I., et al. 2006. Mutations of the mitochondrial holocytochrome c-type synthase in X-linked dominant microphthalmia with linear skin defects syndrome. Am. J. Hum. Genet. 79: 878-889.
5. Wimplinger, I., et al. 2007. HCCS loss-of-function missense mutation in a female with bilateral microphthalmia and sclerocornea: a novel gene for severe ocular malformations? Mol. Vis. 13: 1475-1482.


## CHROMOSOMAL LOCATION

Genetic locus: HCCS (human) mapping to Xp22.2; Hccs (mouse) mapping to X F5.

## SOURCE

HCCS (D-12) is an affinity purified goat polyclonal antibody raised against a peptide mapping near the C-terminus of HCCS of human origin.

## PRODUCT

Each vial contains 200 µg IgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Blocking peptide available for competition studies, sc-168060 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

## STORAGE

Store at 4° C, **DO NOT FREEZE**. Stable for one year from the date of shipment. Non-hazardous. No MSDS required.

## APPLICATIONS

HCCS (D-12) is recommended for detection of HCCS of mouse, rat and human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

HCCS (D-12) is also recommended for detection of HCCS in additional species, including equine, canine, bovine and porcine.

Suitable for use as control antibody for HCCS siRNA (h): sc-91122, HCCS siRNA (m): sc-145904, HCCS shRNA Plasmid (h): sc-91122-SH, HCCS shRNA Plasmid (m): sc-145904-SH, HCCS shRNA (h) Lentiviral Particles: sc-91122-V and HCCS shRNA (m) Lentiviral Particles: sc-145904-V.

Molecular Weight of HCCS: 31 kDa.

## RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use donkey anti-goat IgG-HRP: sc-2020 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible donkey anti-goat IgG-HRP: sc-2033 (dilution range: 1:2000-1:5000), Cruz Marker™ Molecular Weight Standards: sc-2035, TBS Blotto A Blocking Reagent: sc-2333 and Western Blotting Luminol Reagent: sc-2048. 2) Immunofluorescence: use donkey anti-goat IgG-FITC: sc-2024 (dilution range: 1:100-1:400) or donkey anti-goat IgG-TR: sc-2783 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

## RESEARCH USE

For research use only, not for use in diagnostic procedures.

## PROTOCOLS

See our web site at www.scbt.com or our catalog for detailed protocols and support products.